Clinical value of echocardiographic Doppler-derived right ventricular $dp/dt$ in patients with pulmonary arterial hypertension

Koen Ameloot1*, Pieter-Jan Palmers1, Alexander Vande Bruaene1, Annelies Gerits1, Werner Budts1, Jens-Uwe Voigt1†, and Marion Delcroix2†

1Department of Cardiovascular Diseases, Medical Imaging Research Center, University Hospitals Leuven, Leuven, Belgium; and 
2Respiratory Division, University Hospitals Leuven, Leuven, Belgium

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Aims
Right ventricular (RV) $dp/dt$ is the instantaneous rate of RV pressure rise during early systole and is a surrogate marker of RV contractility. The main objective of this study was to evaluate the ability of echocardiographic Doppler obtained RV $dp/dt$ to predict long-term survival in patients with pulmonary arterial hypertension (PAH) and chronic thrombo-embolic pulmonary hypertension (CTEPH).

Methods and results
Seventy-eight consecutive newly diagnosed untreated patients (64 ± 15 years, 71% female, 57% PAH, 43% inoperable CTEPH) were included in the study. At baseline, patients were assessed clinically [New York Heart Association (NYHA) and 6 minutes walking distance (6MWD)], by transthoracic cardiac ultrasound and by right heart catheterization. RV $dp/dt$ was assessed using spectral Doppler recordings from the tricuspid regurgitation signal at a sweep speed of 200 mm/s by measuring the time interval in which the regurgitant velocity increased from 0.5 to 2 m/s. During a mean follow-up period of 3.5 ± 1.7 years, 31 patients died and 3 received a lung transplant [study endpoint reached in 34/78 (44%) patients]. The optimal RV $dp/dt$ cut-off was determined by receiver operating characteristic analysis at 3 years to be 410 mmHg/s (specificity 84%, positive-predictive value 55%, and negative-predictive value 83%). In univariate analysis, RV $dp/dt$, 410 mmHg/s (hazard ratio 2.67, 95% CI 1.30–5.47, $P$ = 0.007), tricuspid annulus plane systolic excursion (TAPSE), NYHA, 6MWD, and right atrial pressure were predictors of mortality. In a multivariate model with TAPSE, RV $dp/dt$ remained an independent predictor of mortality ($P$ = 0.01).

Conclusion
A reduced baseline RV $dp/dt$ is a clear indicator of poor outcome independent of TAPSE in patients with PAH/CTEPH.

Keywords
Right ventricular function • Pulmonary hypertension

Introduction
Adequate assessment of right ventricular (RV) function is an essential component of risk stratification in patients with pulmonary hypertension (PH) since two-thirds of total mortality may be attributed to RV failure.1,2 The complex 3D shape of the RV and poor endocardial definition greatly complicate volumetric RV function techniques by 2D echocardiography.3 The ideal RV function parameter should predict long-term clinical outcome, be easy to obtain, reproducible, and parallel functional capacity. Current guidelines recommend the use of a combination of clinical [New York Heart Association (NYHA) and 6 minutes walking distance (6MWD)], invasive [right atrial pressure (RAP) and cardiac index (CI)], and echocardiographic parameters for risk stratification in PH patients. Many echocardiographic parameters are related to prognosis in PH.4–11 However, awaiting further validation studies, only TAPSE and the presence of pericardial fluid are recommended in the guidelines as prognostic parameters in PH.12

RV $dp/dt$ is the instantaneous rate of RV pressure rise during early systole and is a surrogate marker of RV contractility. RV $dp/dt$ can easily be obtained by continuous-wave Doppler of the tricuspid regurgitation (TR) jet and measuring the time interval necessary for...
the RV to build up a predefined pressure difference. Non-invasive Doppler obtained RV peak positive dp/dt showed a good correlation with right heart catheterization (RHC)-derived dp/dt, irrespective of the level of RAP.\textsuperscript{13,14} There are no data on the value of RV dp/dt as a predictor of long-term clinical outcome. Therefore, we performed this validation study to evaluate (i) the ability of RV dp/dt to predict long-term survival in patients with pulmonary arterial hypertension (PAH) and chronic thrombo-embolic pulmonary hypertension (CTEPH), (ii) its correlation with functional capacity, and (iii) interobserver variability.

**Methods**

**Study design**

All patients with PH referred to our tertiary care hospital are consecutively included in the ‘UZ Leuven PH database’. For this study, we selected all newly diagnosed and so far untreated adult patients referred between 2006 and 2009 with PAH or inoperable CTEPH.\textsuperscript{15} Baseline assessment includes clinical parameters (NYHA and 6MWD if feasible), echocardiography, and RHC. Eight patients were excluded because of an inappropriate TR signal. The final study population consisted of 78 patients. All patients received appropriate state-of-the-art treatment including phosphodiesterase-5 inhibitors, endothelin receptor antagonists, pros-tacyclin analogues, or atrial septostomy. Primary endpoint was all-cause mortality. Three patients with irredeemable deterioration received a lung transplant and were assimilated to death. Survival time was estimated as the time from baseline echocardiography to the end of the study (31 December 2012) or death/transplantation. At study termination, the vital status was checked by review of the medical records or by phone contact. A minimal follow-up period was 3 years. The follow-up rate was 100%. The ethics committee of UZ Leuven approved the protocol. Because the protocol was judged to pose a low risk, the obligation for a written informed consent was waived.

**Echocardiography**

Baseline echocardiographies were performed on a Vivid 7 or Vivid E9 ultrasound system (GE Vingmed Ultrasound, Horten, Norway) as well as Philips iE 33 (Royal Philips Electronics, Eindhoven, Netherlands). Baseline recordings were retrospectively re-analysed using a clinical post-processing workstation (EchoPAC, GE Vingmed Ultrasound) by two completely blinded cardiologists (Ameloot Koen and Palmers PieterJan). Both were not involved in routine assessment of study patients and were unaware of the clinical course of patients during follow-up. RV dp/dt was assessed using spectral Doppler recordings from the TR signal at a sweep speed of 200 mm/s by measuring the time interval in which the regurgitant velocity increased from 0.5 to 2 m/s (1–16 mmHg) (Figure 1). These velocities were chosen as being most robust for reading while resulting in the best correlation with invasive measurements.\textsuperscript{15} In patients with sinus rhythm, three random beats were averaged to obtain the final RV dp/dt value. In six patients with atrial fibrillation, five beats were averaged. Furthermore, a complete 2D, Doppler, and M-mode examination was performed as recommended in guidelines for the assessment of PH patients.\textsuperscript{12,16}

**Haemodynamics**

In 49 patients, routine RHC with the measurement of pulmonary pressures, mixed venous oxygen saturation, and assessment of Cl by thermodilution and Fick’s principle was performed within 10 days of baseline echocardiography. In 21 patients, RHC was performed with a larger time interval (median 26 days). Their data were not used for correlation with echocardiographic data. One PAH patient refused RHC. In seven CTEPH patients, with mild PH and preserved functionality (NYHA I) or with advanced age or co-morbidity, no RHC was performed. None of these patients had echocardiographic evidence of left-sided diastolic dysfunction.

**Data analysis**

Continuous variables were summarized as mean ± SD or as median with interquartile range as appropriate, and categorical variables as the number of subjects and percentage. A predictive model for all-cause mortality was constructed. First, a receiver operating characteristic (ROC) curve was constructed to test the ability of RV dp/dt to predict mortality within 3 years after baseline echocardiography. Secondly, uni- and multivariate Cox proportional hazards regression models were used to describe the prognostic properties of RV dp/dt in relationship with the other clinical, haemodynamic, and echocardiographic parameters. A two-step multivariate model was used: first, RV dp/dt was adjusted for TAPSE to construct a model with only echocardiographic predictors. Secondly, the combination of these echocardiographic predictors was adjusted for clinical parameters (NYHA and 6MWD) to construct a model with only non-invasive parameters. Thirdly, survival curves using the Kaplan–Meier method were constructed and compared by the log-rank test. Finally, baseline and ‘final’ RV dp/dt were compared by a paired-sample Wilcoxon signed-rank test in patients who died during follow-up if a final echo was available < 6 months before death. Patients surviving < 6 months were excluded from this analysis. Subgroups with preserved and reduced RV dp/dt were compared by unpaired Student’s t-tests. To assess the relation between RV dp/dt and functional capacity, ANOVA was used to compare the proportion of patients with preserved RV dp/dt per NYHA class and linear regression to describe correlation with 6MWD. Interobserver variability was assessed by linear regression, Pearson correlation coefficients, Bland–Altmann plot, and by the percentage interobserver variability (absolute difference between two observations divided by the means and expressed as percentage). Data by both observers were compared using paired-sample Student’s t-test. The level of concordance was calculated as the proportion of patients classified similarly by both observers (either preserved or reduced RV dp/dt). Statistical analysis was performed using the Matlab software (version R2010b, Mathworks, USA). A P-value of <0.05 was considered significant.

**Results**

**Study population**

Baseline parameters illustrate the disease severity of the study cohort (Table 1). During a mean follow-up period of 3.5 ± 1.7 years (minimal 3.0–maximal 6.7 years), 31 patients died (mean after 2.3 ± 1.6 years) and 3 received a lung transplant. The study endpoint was reached in 34/78 (44%) of the patients. The vast majority of the study patients died due to a combination of respiratory insufficiency and right-sided heart failure with refractory fluid overload or arrhythmias.

**Survival analysis**

A ROC curve on the ability to predict all-cause mortality within 3 years after echocardiography identified the optimal cut-off for RV dp/dt to be 410 mmHg/s (specificity 84%, positive-predictive value 55%, and negative-predictive value 83%) (Figure 2). Efficiency (the proportion of patients correctly classified) was 76%. RV dp/dt, TAPSE, 6MWD, and NT-proBNP showed good and equivalent
The current study shows for the first time that a reduced baseline RV dp/dt was associated with a higher NYHA class and lower 6MWD (Table 1). The proportion of patients with a preserved RV dp/dt was significantly lower with an increasing NYHA class (Figure 5). Linear regression revealed a weak significant correlation between RV dp/dt and 6MWD (R = 0.28, P = 0.02).

Interobserver variability

The level of concordance between observers regarding the distinction of ‘preserved’ and reduced RV dp/dt was high (92.3%). Interobserver variability was 9.9%. Pearson correlation coefficient was 0.63, with a mean bias 59.9 mmHg/s. The Bland–Altman plot revealed that most of this variability was attributable to measurement variation in patients with a high (i.e. preserved) RV dp/dt. A paired-sample t-test did not show significant differences between datasets by both observers (P = 0.10; Figure 6).

Discussion

Survival analysis

The current study shows for the first time that a reduced baseline RV dp/dt as a surrogate marker for reduced RV contractility is a clear indicator of poor prognosis in patients with PAH/CTEPH. A previous
study showed the mean echocardiographic RV dp/dt in healthy subjects to be 1016 mmHg/s.\textsuperscript{17} This contrasts with our presented RV dp/dt cut-off (<410 mmHg/s) and with the mean RV dp/dt in our study cohort (658 ± 371 mmHg/s). A reduced baseline RV dp/dt was associated with a 2.57-fold increased risk of death and a 3-year survival rate of 45% compared with 83% in patients with a preserved baseline RV dp/dt. We also showed that RV dp/dt is a dynamical parameter that further decreases in patients short before death. The prognostic properties of RV dp/dt have not been evaluated before. Therefore, routine determination of RV dp/dt is currently not recommended in the guidelines.\textsuperscript{12,16} In contrast, TAPSE has been extensively validated in previous studies and is widely used as the most accurate echocardiographic tool for risk stratification in patients with PH- or non-PH-induced RV failure.\textsuperscript{4–6,11} We demonstrated that RV dp/dt and TAPSE have equivalent accuracy in predicting long-term survival and are additive to each other. Three major physiological explanations for these findings apply. First, RV dp/dt reflects the complex interaction of all muscle fibres building up RV pressure during early

<table>
<thead>
<tr>
<th>Demographics</th>
<th>All patients</th>
<th>RV dp/dt (mmHg/s)</th>
<th>P-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of patients</td>
<td>78</td>
<td>21</td>
<td>57</td>
</tr>
<tr>
<td>Age (years)</td>
<td>63.6 ± 15.2</td>
<td>69.8 ± 14.7</td>
<td>61.4 ± 14.9</td>
</tr>
<tr>
<td>Gender (female), n (%)</td>
<td>55 (70%)</td>
<td>12 (57%)</td>
<td>43 (75%)</td>
</tr>
<tr>
<td>Cause pulmonary hypertension, n (%)</td>
<td>CTEPH 34 (43%)</td>
<td>8 (38%)</td>
<td>26 (46%)</td>
</tr>
<tr>
<td>PAH 44 (56%)</td>
<td>13 (62%)</td>
<td>31 (54%)</td>
<td></td>
</tr>
<tr>
<td>Clinical characteristics [n = 78 (NYHA), n = 68 (6MWD), n = 49 (NT-proBNP)]</td>
<td>NYHA</td>
<td>2.8 ± 0.7</td>
<td>3.1 ± 0.6</td>
</tr>
<tr>
<td>6MWD (m)</td>
<td>309 ± 160</td>
<td>259 ± 122</td>
<td>356 ± 149</td>
</tr>
<tr>
<td>NT-proBNP (ng/L)</td>
<td>2059 (635; 4439)</td>
<td>3085 (695; 6217)</td>
<td>1877 (630; 3450)</td>
</tr>
</tbody>
</table>

| Right heart haemodynamics (n = 70, 49 patients within 10 days of echo) | Heart rate (bpm) | 77.7 ± 16.0 | 75.6 ± 13.3 | 80.3 ± 16.0 |
| RAP (mmHg) | 5 (3; 9) | 4 (3; 13) | 5 (2.2; 10.7) |
| sPAP (mmHg) | 77.6 ± 20.0 | 83.3 ± 21.3 | 75.9 ± 21.4 |
| dPAP (mmHg) | 29.4 ± 10.6 | 34.2 ± 13.7 | 29.0 ± 10.1 |
| mPAP (mmHg) | 44.7 ± 11.8 | 48.9 ± 11.4 | 43.5 ± 12.7 |
| PAOP (mmHg) | 8.1 ± 3.9 | 8.9 ± 4.4 | 7.9 ± 4.1 |
| CI (L/min/m²) | 2.0 ± 0.5 | 2.0 ± 0.5 | 2.0 ± 0.6 |
| SVI (mL/m²) | 280 ± 9.4 | 24.5 ± 7.0 | 28.0 ± 9.4 |
| SVO₂ (%) | 62.3 ± 7.7 | 59.9 ± 5.3 | 63.1 ± 7.4 |
| PVRI (dyne s cm⁻⁻) | 891 ± 409 | 919 ± 424 | 914 ± 458 |

| Echocardiographic parameters (n = 78) | RVEDA (cm²) | 28.0 ± 7.9 | 31.3 ± 7.6 | 26.8 ± 7.7 |
| RVEDA/LVEDA | 1.5 ± 0.8 | 1.9 ± 1.0 | 1.3 ± 0.6 |
| LVEF (%) | 64.0 ± 7.1 | 63.4 ± 8.0 | 64.2 ± 6.8 |
| Grade TR | 2.7 ± 0.8 | 2.9 ± 0.8 | 2.6 ± 0.9 |
| sPAP (mmHg) | 70.8 ± 19.0 | 78.3 ± 18.7 | 68.1 ± 18.5 |
| PAT (ms) | 70.6 ± 20.3 | 61.5 ± 21.7 | 74.3 ± 18.8 |
| RVET (ms) | 297 ± 68 | 290 ± 56 | 300 ± 72 |
| IVC collapsibility, n (%) | 50 (64%) | 16 (76%) | 34 (59%) |
| Pericardial fluid, n (%) | 25 (32%) | 9 (43%) | 16 (29%) |
| TAPSE (mm) | 17 ± 5 | 16 ± 5 | 17 ± 6 |
| TEI | 0.7 ± 0.3 | 0.8 ± 0.3 | 0.6 ± 0.3 |

CTEPH, chronic thromboembolic pulmonary hypertension; PAH, pulmonary arterial hypertension; NYHA, New York Heart Association; 6MWD, 6 minutes walking distance; RAP, right atrial pressure; sPAP, systolic pulmonary arterial pressure; dPAP, diastolic pulmonary arterial pressure; mPAP, mean pulmonary arterial pressure; PAOP, pulmonary arterial occlusion pressure; CI, cardiac index; SVI, stroke volume index; SVO₂, mixed venous oxygen saturation; PVRI, pulmonary vascular resistance index; RVEDA, right ventricular end-diastolic area; LVEDA, left ventricular end-diastolic area; LVET, left ventricular ejection fraction; TR, tricuspid regurgitation; PAT, pulmonary acceleration time; RVET, right ventricular ejection time; IVC, inferior vena cava; TAPSE, tricuspid annulus plane systolic excursion.

Table 1  Baseline demographics, diagnosis, clinical characteristics, haemodynamic and echocardiographic parameters of the overall cohort and stratified by normal or reduced RV dp/dt
systole, whereas TAPSE predominantly reflects shortening of the longitudinal oriented muscle fibres primarily involved in ejection of blood.\textsuperscript{3,18} Other surrogate parameters of isovolumetric contraction such as tissue Doppler obtained peak isovolumetric contraction velocity also showed to be a predictor of mortality in PH patients.\textsuperscript{19} It may be hypothesized that a steeper rate of RV pressure rise during early systole results in an earlier pulmonary valve opening, thereby extending ejection time and increasing RV stroke volume.

Secondly, many patients with severe PH also have moderate or severe TR (57% in our study population). In patients with severe TR, TAPSE represents the combination of forward and backward flow and may be too optimistic as an estimator of true forward RV stroke volume. However, accuracy of RV dp/dt in these patients may also be questioned since severe TR results in faster RA–RV pressure equilibration. Moreover, the usually neglected inertia term of the complete Bernoulli equation may become relevant in severe TR.

Thirdly, TAPSE can be falsely elevated in patients with a failing RV

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**Figure 2:** ROCs on the ability of RV dp/dt to predict all-cause mortality within 3 years after baseline echocardiography. The arrow indicates the optimal cut-off point (RV dp/dt 410 mmHg/s). AUC, area under curve.

**Table 2** Area under the ROC curve, sensitivity, specificity, positive- and negative-predictive values, and efficiency for suggested cut-off points for RV dp/dt and other echocardiographic, clinical, biochemical, and RHC parameters to predict all-cause mortality within 3 years after baseline echocardiography

<table>
<thead>
<tr>
<th>Parameter</th>
<th>RV dp/dt</th>
<th>TAPSE</th>
<th>Tei</th>
<th>6MWD</th>
<th>proBNP</th>
<th>RAP</th>
<th>CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>AUC</td>
<td>0.67</td>
<td>0.66</td>
<td>0.50</td>
<td>0.75</td>
<td>0.79</td>
<td>0.50</td>
<td>0.61</td>
</tr>
<tr>
<td>95% CI</td>
<td>0.53–0.81</td>
<td>0.51–0.80</td>
<td>0.36–0.65</td>
<td>0.60–0.89</td>
<td>0.64–0.96</td>
<td>0.35–0.65</td>
<td>0.46–0.76</td>
</tr>
<tr>
<td>P-value</td>
<td>0.01</td>
<td>0.01</td>
<td>NS</td>
<td>0.0003</td>
<td>0.0001</td>
<td>NS</td>
<td>NS</td>
</tr>
<tr>
<td>Optimal cut-off</td>
<td>&lt;410 mmHg/s</td>
<td>&lt;15 mm</td>
<td>&gt;0.70</td>
<td>&lt;200 m</td>
<td>&gt;1400 ng/L</td>
<td>&lt;10 mmHg</td>
<td>&lt;2.0 L/min/m²</td>
</tr>
<tr>
<td>Sensitivity</td>
<td>52.38</td>
<td>52.38</td>
<td>52.38</td>
<td>55.56</td>
<td>83.33</td>
<td>31.58</td>
<td>70.00</td>
</tr>
<tr>
<td>Specificity</td>
<td>84.21</td>
<td>75.00</td>
<td>44.64</td>
<td>86.00</td>
<td>75.68</td>
<td>88.00</td>
<td>55.10</td>
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<tr>
<td>PPV</td>
<td>55.00</td>
<td>44.00</td>
<td>26.19</td>
<td>58.82</td>
<td>52.63</td>
<td>50.00</td>
<td>38.89</td>
</tr>
<tr>
<td>NPV</td>
<td>82.76</td>
<td>80.77</td>
<td>71.43</td>
<td>84.31</td>
<td>93.33</td>
<td>77.19</td>
<td>81.82</td>
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<tr>
<td>Efficiency</td>
<td>75.64</td>
<td>68.83</td>
<td>46.75</td>
<td>77.94</td>
<td>77.55</td>
<td>72.46</td>
<td>59.42</td>
</tr>
<tr>
<td>PLR</td>
<td>3.32</td>
<td>2.10</td>
<td>0.95</td>
<td>3.97</td>
<td>3.32</td>
<td>2.58</td>
<td>1.56</td>
</tr>
</tbody>
</table>

TAPSE, tricuspid annulus plane systolic excursion; 6MWD, 6 minutes walking distance; BNP, brain natriuretic peptide; CI, cardiac index; AUC, area under curve; 95% CI, 95% confidence interval; PPV, positive-predictive value; NPV, negative-predictive value.
Our data confirm the validity of NYHA, 6MWD, and RAP as indicators of long-term prognosis in PH. Compared with these predictors, RV dp/dt showed equivalent ROC accuracy and an equally significant hazard ratio (HR). However, our data suggest an added value of RV dp/dt. We therefore propose an echocardiographic model combining TAPSE and RV dp/dt. This model remained an independent predictor of mortality after adjusting for NYHA and 6MWD.

Of note, RAP was associated with a significant HR, but ROC analysis could not demonstrate a significant accuracy to predict 3-year mortality. Indeed, patients with an elevated baseline RAP probably have more profound RV failure and a worse shorter-term prognosis.

Echocardiographic parameters such as Tei index, presence of pericardial effusion, and TR grade were no significant predictors of mortality in our study. Besides that, in previous studies, these parameters did not add prognostic information independent of TAPSE, 6MWD, or NYHA class. Conflicting results with previous studies might be explained by marked differences in the study population (disease severity, treatment status, and types of PH) and study endpoints used. Finally, we did not find major differences with respect to pulmonary pressures or CI when patients were stratified according to normal or reduced RV dp/dt. Indeed, it has been shown previously that invasive hemodynamics correlate with exercise capacity, but are poorly predictive for adverse clinical outcome.

### Functional capacity

Our data showed that the proportion of patients with a preserved RV dp/dt was inversely related to NYHA class. Also, RV dp/dt correlated significantly with 6MWD. These results are in line with previous studies showing correlation between RV dp/dt and functional NYHA class in patients with mitral stenosis and also between RV dp/dt and pulmonary pressures or CI when patients were stratified according to NYHA class. It is known that the relationship between RV functional parameters and exercise capacity is not linear in PH due to confounding factors such as impaired pulmonary function, deconditioning, and presence of systemic illness limiting exercise capacity irrespective of RV function. Therefore, previous studies failed to validate a decreased 6MWD as a surrogate for clinical endpoints in patients with RV failure. Moreover, many RV function parameters including TAPSE have been shown to be loading dependent. However, our attempt to correct for loading by indexation of RV dp/dt to pulmonary pressure or RVEDA did not improve prognostic properties or correlation with functional parameters.

### Interobserver variability

We could demonstrate a high level of concordance between observers when RV dp/dt was used as a dichotomous parameter. The percentage interobserver variability was equal to values previously reported for TAPSE. The Bland and Altman plot clearly illustrates that the rather low correlation coefficient was mainly driven by two outliers and by variability in the high normal range of the parameter. This may be because the time measurement in the RV dp/dt denominator becomes less reliable for shorter time intervals, i.e. higher RV dp/dt values. Recalculating Pearson correlation coefficient by excluding two outliers reveals a $R$-value of 0.81, which is in between values previously reported for TAPSE ($R = 0.94$) and right ventricular fractional area change ($R = 0.64$). In summary, discrimination between a preserved and reduced RV dp/dt is easy and reproducible.
However, it is difficult to differentiate within the preserved subgroup how good RV contractility actually is. We therefore recommend using RV $dp/dt$ as a dichotomous parameter.

**Strengths—limitations—future perspectives**

This is, to our knowledge, the largest study in the modern treatment era investigating indicators of prognosis exclusively in newly diagnosed and so far untreated PH patients. The median delay between first diagnosis and baseline ultrasound was only 62 days. This prevented blurring of our results by inclusion of long-term stable patients, which is a major bias of many previous studies in PH. The reliability of the time-to-event analysis was further reinforced by the prolonged minimal follow-up of 3 years with a corresponding mortality rate of 40%. This is in agreement with values previously reported and allowed the use of all-cause mortality as the primary study endpoint. The use of a prospective, high-quality database previously used in several studies offers excellent protection against selection...
bias due to incompleteness of the database. The retrospective re-
analyses of the images excludes the theoretical possibility that
knowledge on baseline RV dP/dt values might have influenced base-
line assessment or clinical course of the patients.

A first limitation is that our study population might be enriched by
selection bias due to exclusion of eight patients without significant
TR. In previous studies, patients with severe TR had a worse prog-
nosis. We also excluded operable CTEPH patients since their
prognosis is mainly determined by the success of the intervention.
Inoperable patients are generally older, have more co-morbidity,
and have a worse prognosis. This raises the question whether the
prognostic importance of RV dP/dt would hold among a less ill pop-
ulation. Secondly, we did not take RAP in to account to determine
RV dP/dt. This is because a previous study found RV dP/dt to be
independent of RAP and because inclusion of RAP would greatly
complicate RV dP/dt assessment and extend the time needed for
its calculation. A final remark is that only patients with PAH/
CTEPH were included. Future studies are necessary to determine
whether our results can be extrapolated to patients with other
types of PH- or non-PH-induced RV failure.

Conclusion

RV dP/dt is easy to obtain and reproducible when used as a dichotom-
ous parameter. A reduced baseline RV dP/dt as a surrogate marker
of reduced RV contractility is an independent indicator of poor
outcome in patients with PAH/CTEPH. Baseline RV dP/dt parallels
functional capacity as assessed by NYHA and 6MWD. RV dP/dt can
be used with TAPSE, NYHA, and 6MWD for complete non-invasive
risk assessment in patients with PH.

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An adolescent with laminopathy presenting as ventricular tachycardia and left ventricular apical aneurysm

Hye Won Kwon, Bo Sang Kwon*, Eun Jung Bae, and Jong Hee Chae

Department of Pediatrics, Seoul National University Children’s Hospital, 101 Daehak-ro, Jongno-gu, Seoul 110-744, South Korea

* Corresponding author. Tel: +82 2 2072 4193; Fax: +82 2 743 3455, E-mail: ciec10@nmu.ac.kr

A 17-year-old girl was admitted because of recurrent ventricular tachycardia after sudden cardiac arrest (Panel A). Two-dimensional and strain echocardiography revealed a dilated left ventricle (LV) with apical aneurysm and dyskinesia (Panels B and C, and see Supplementary data online, Video S1). The normal coronary artery was confirmed under coronary angiography (Panel D, and see Supplementary data online, Videos S2 and S3). Cardiac magnetic resonance imaging revealed apical aneurysmal dilatation and wall thinning with thrombus in the LV (Panel E and see Supplementary data online, Video S4). Subendocardial and transmural late gadolinium enhancement in the LV apical and lateral walls was evident (Panel F, arrow heads). An implantable cardioverter-defibrillator was applied in addition to amiodarone, enalapril, carvedilol, and warfarin therapies.

The patient complained of a slowly progressive lower extremity weakness over the past 10 years. She showed a waddling gait, Gower’s sign, and Achilles tendon contracture. Under the impression of cardiomyopathy associated with muscular dystrophy, we performed genetic testing, which revealed a mutation (c.1621C>A [p.Arg541Ser]) in the LMNA gene.

This is an unusual case of laminopathy presenting as ventricular tachycardia associated with localized myocardial fibrosis and aneurysm.

Supplementary data are available at European Heart Journal — Cardiovascular Imaging online.

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